MR Tumor Staging for Treatment Decision in Case of Wilms Tumor

G. Schneider, M.D., Ph.D.; P. Fries, M.D.

Dept. of Diagnostic and Interventional Radiology, Saarland University Hospital, Homburg/Saar, Germany

Introduction

Nephroblastoma – also known as Wilms tumor – is the most frequent renal malignancy in childhood with the highest incidence of this tumor within the fourth year of life. 80% of patients are less than 5 years old, however it is a rare condition in neonates (<1%). In general, there are no known risk factors for the development of nephroblastoma, but it may be associated with rare conditions like Denys-Drash (triad of congenital nephropathy, Wilms tumor and intersex disorders), WAGR (also called Wilms tumor-aniridia syndrome) and Beckwith-Wiedeman (giantism associated with tumors and malformations) syndrome. The incidence is approx. 1: 100,000 for western countries including the US, while a lower incidence is reported for Asian countries. If not associated with a syndrome, clinical symptoms – if present at all – are very often unspecific and abdominal pain and palpable tumor can be the only findings at the time of diagnosis. MRI is considered the imaging modality of choice for tumor staging and subsequent treatment planning. If imaging is conclusive, often no biopsy is performed prior to initiation of therapy. Clinical treatment is according to protocols of SIOP (Society of Pediatric Oncology) in Europe or COG (Children’s Oncology Group) in North America. Therapy includes primary surgery (COG), preoperative chemotherapy (SIOP), and/or adjuvant chemotherapy. If not treated, prognosis of a Wilms tumor is poor. Independent of prognostic factors such as stage and grading, the overall outcome is good and approximately 90% of all children will be cured. Questions for imaging are: a) supporting the suspicion of a Wilms tumor for initiation of therapy, b) evaluation of tumor volume, c) contralateral tumor manifestation and d) lymph nodes metastasis or infiltration of neighboring structures e.g. diaphragm or liver.

Tumor staging has to include at least the whole abdomen and thorax (lung filiae are the most common presentation of metastatic disease). Imaging modalities used are ultrasound, MRI, and CT in case of lung metastases. Depending on final tumor histology, a bone (often scintigram) and brain MRI scan have to be performed in case of CCSK (clear cell sarcoma) and RTK (rhabdoid tumor of the kidney), too. MRI is recommended independent of the above-mentioned reasons in any case where a) a caval vein tumor thrombus, b) infiltration of liver and diaphragm, or c) continuous tumor extension into the thoracic cavity is suspected.

Patient history

A 4-year-old girl presented with a large palpable mass in the left upper quadrant and unspecific abdominal pain. Ultrasound had already revealed a large tumor of the left hemiabdomen with mass effect towards the liver. The patient was referred to our MRI department because of suspicion of Wilms tumor.

MRI protocol

MRI was conducted using a 1.5 Tesla MAGNETOM Aera with the combination of the 18-channel body coil and the integrated spine coil. For the MRI procedure the patient received an intravenous sedation using propofol. The imaging protocol included diffusion-weighted imaging (DWI, syngo REVEAL), acquired during free breathing, and transversal T2w TSE and HASTE sequences with navigator triggering. A single-shot echo planar diffusion imaging with Stejskal-Tanner diffusion encoding scheme was applied. For fat saturation, an inversion recovery technique was used. The sequence parameters were:

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Transversal high-resolution T2w images showing the Wilms tumor (*) and multiple lung metastases (arrows). Due to the space occupying aspect of the large tumor, the residual kidney is swollen (+) and also slight edema of the liver hilum can be seen (arrowheads).
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2 Rotating MIP based on high b-value images.

3 (A) Coronal DWI MIP. Original b-value images at 0 and 800 s/mm² (B, C and E, F) as well as calculated b-value at b 1400 s/mm² (D, G) are shown. (Arrows pointing to lung metastases.)

4 Calculated ADC map (A) and corresponding T2w image (B) demonstrating the tumor heterogeneity. The area marked by the arrows has a clear restriction in diffusibility but based on T2w imaging alone, no differentiation between this area and the one marked with * is possible. While the high signal area on T2w and high ADC values may represent cysts or calceal dilation, the area with the high restriction of diffusion represents a very densely packed areal e.g. mucous tumor cells.
Based on ADC maps and high b-value images (b 1400 s/mm² is shown), a clear differentiation between residual but swollen kidney tissue (arrows) and the Wilms tumor (arrowhead) is possible. Both types of tissue differ in their cellular density, however, on T2w images no clear differentiation is possible in this case (compare Fig. 1). Nevertheless, not all areas of the tumor are characterized by high signal on the very high b-value images, demonstrating well the tumor heterogeneity. (A, B) ADC maps. (C, D, E) b 1400 s/mm² images. (F) Coronal thick-slice MPR based on b 1400 s/mm² images (* spleen).
TR 15400 ms, TE 75 ms, TI 180 ms, PAT factor of 2, 3-scan trace (averaged), FOV 309 x 380, matrix 208 x 128 (interpolated to 208 x 256), slice thickness 5 mm, no gap, 4 averages. Real voxel size was 1.5 x 3 x 5 mm³. Two b-values at b 0 and b 800 s/mm² were acquired. ADC maps and additional high b-value images at b 1400 s/mm² were calculated automatically by the scanner software, based on linear signal decay. DWI covered the whole body trunk from skull base towards upper lower extremities. Acquisition time was approx. 15 min. For presentation and fast overview about tumor spread, a rotating maximum intensity projection (MIP) based on b 800 s/mm² was generated.

For detailed morphology and assessment of tumor infiltration, navigator triggered T2w TSE was applied for the abdomen including the lower thorax and mediastinum. Sequence parameters were TR 3508 ms, TE 102 ms, 2 averages. PAT factor 2, FOV 188 x 250 mm², matrix 269 x 512, slice thickness 6 mm, 20% gap, acquisition time was approx. 8 min.

**Imaging findings**

A large occupying tumor deriving from the lower pole of the left kidney with compression of the residual kidney and mass effect towards the liver and especially the left liver lobe is shown. Due to the mass effect, slight edema of the liver hilus can be seen. However, the border of the mass is well circumscribed and no evidence of diffuse tumor infiltration of the liver, spleen or diaphragm can be seen. Since no encasement of retroperitoneal vessels or other structures is seen DD of neuroblastoma can be ruled out. Also the lumen of the abdominal aorta is regular and neither a tumor infiltration of the large vessels nor a tumor-thrombus can be visualized. The right kidney and the other abdominal organs are free of metastases. However, already well visualized by the MIP DWI, a large tumor mass at the right lung hilum can be seen with compression of central lung structures and edema of the depending lung tissue. In addition, at least four additional lung metastases are detected. No evidence for bone metastases. The bright signal of the bone marrow on high b-value images has to be considered as age related. The size of the displaced spleen is also within normal age-related range.

On a follow-up study after chemotherapy and before surgery a tremendous reduction of tumor size can be noticed. Only small residual tumor tissue of one lung metastases is visible on CT and MRI.

**Conclusion**

Whole-body imaging in staging of Wilms tumor can replace CT imaging and gives all necessary information for therapy planning. With the help of newer imaging modalities in MRI, especially DWI, the prediction of tumor response needs to be evaluated. This can easily be done by correlating histological data with imaging data from patients enrolled in prospective clinical trials. As preoperative chemotherapy is only part of the SIOP studies such investigations can predominantly be performed in Europe.
Follow-up examination with CT (A) still showing a small residual lung metastasis (arrow), which can also be visualized by MRI (B). The main tumor is also clearly reduced in its mass after first cycle of chemotherapy (C–F) in caudo-cranial sorting.

References

Contact
PD Dr. Dr. Günther Schneider
Dept. of Diagnostic and Interventional Radiology
Saarland University Hospital
Kirberger Strasse 66421 Homburg/Saar
Germany
dr.guenther.schneider@uks.eu